Short Time Recurrence of Retroperitoneal Desmoid Tumor in Patient With Familial Adenomatous Polyposis

Danyal O Imam MBBS; Steve Serrao, MD; Christopher Bent, MD; Ronaldo D.Gnass, MD; Pejman Solaimani, MD

Introduction

A Desmoid Tumor (DT) is a locally invasive, non-metastatic mesenchymal stem cell tumor. It typically manifests as a soft tissue mass occurring in muscles, fasciae and aponeuroses.

Case Description

A 34 year old gentleman presented with abdominal pain

↓

CT abdomen demonstrated a large (11 x 14.4 x 14 cm) intrabdominal mass

↓

Transcatheter biopsy of the mass confirmed DT

↓

Bidirectional endoscopy demonstrated greater than a hundred adenomatous colon polyps and genetic testing confirmed APC mutation.

↓

Diagnosed with DT associate with Familial Adenomatous Polyposis (FAP)

↓

Radical resection of mesenteric mass and small bowel (20 cm of terminal ileum), omentectomy as well as total colectomy with ileorectal anastomosis.

↑

Three months later, he was re-admitted for increased abdominal girth with abdominal pain.

↑

MRI scans demonstrated a new, extensive, lobulated soft tissue mass occupying the entire abdominal cavity

↑

Biopsy of the mass confirmed recurrent DT.

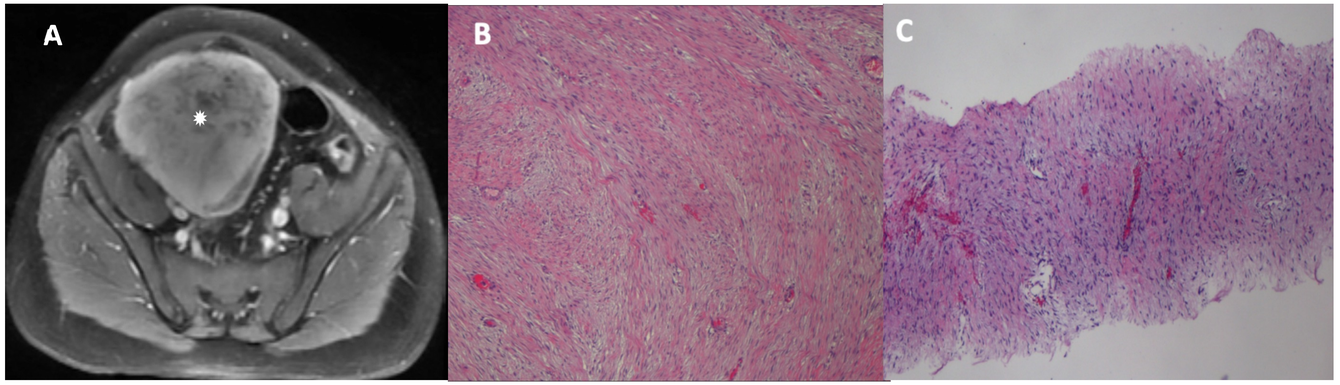
↑

Due to the large size and proximity to the superior mesenteric artery, the mass was deemed to be unresectable.

↑

Patient was started on palliative systemic treatment with liposomal Doxorubicin, however, due to lack of response, he was then switched to weekly methotrexate and Vinblastine.

Imaging and Pathology



A: T1 weighted post contrast MR axial image through the pelvis demonstrating large enhancing right pelvic extraperitoneal soft tissue mass (white star); B: Spindle cell lesion, with fascicular and loose storiform growth patterns (H&E, 40x) prior to resection; C: Recurrent tumor with pathology demonstrating spindle cell lesion with more edema and/or myxoid stroma.

Discussion

- DTs are rare soft tissue neoplasms that make up up only 0.03% of all neoplasms and 3% of soft tissue tumors. They typically involve the abdominal wall, extremities, and mesentery.
- Although the etiology of DTs is not well known, there are several risk factors that are associated with their occurrence including: genetic syndromes such as FAP and Gardner syndrome, antecedent trauma to the site of the lesion, pregnancy, and female sex.
- Numerous studies have been done assessing the efficacy of surgery, radiotherapy, and chemotherapy in managing DTs. However, none have shown conclusive benefit.
- The recurrence rate, particularly following surgical management, has been found to be as high as 77%.
- Given the lack of definitive management and high recurrence rates, treatment options should likely include combination of both surgical intervention and systemic treatment after discussing options and adverse effects of treatment with the patient.